

Uncommon presentation of Kimura's disease –a case report

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Abstract: Kimura's disease is a chronic inflammatory disorder of unknown origin usually seen in adult males. The most common clinical feature of this disease is an asymptomatic unilateral soft-tissue mass in the head and neck, frequently involving major salivary glands and lymph nodes. Bilateral involvement is rare. We herein report an uncommon presentation of this disease where an elderly female presented with bilateral auricular masses with multiple swellings in head and neck. Treated medically with steroids and antihistamine and no recurrence of the disease on follow up till date.

I. Introduction

Kimura's disease is a chronic inflammatory disorder of unknown origin usually seen in adult males (1–8). The most common clinical feature of this disease is an asymptomatic unilateral soft-tissue mass in the head and neck, frequently involving major salivary glands and lymph nodes(1). Bilateral involvement is rare. We herein report an uncommon presentation of this disease where an elderly female presented with bilateral auricular masses with multiple swellings in head and neck.

II. Case report:

An elderly (75yr) female presented with bilateral multiple neck swellings since two months, difficulty in swallowing since one week. On examination, up to five multiple swellings in the left side, distributed in preauricular, sub mental, submandibular and upper deep cervical region were noted, all swelling ranged from 2-3cm in size firm, non-tender, non pulsatile with smooth surface and rounded border(figure-1). Similarly on the Right side preauricular lymph node was enlarged, measuring 3x3cm in size, with similar features as noted on the other sides. Indirect laryngoscopy shows congested and edematous epiglottis, bilateral aryepiglottic fold, bilateral vocal cord were normal(figure 3-4). Nose and PNS examination shows DNS to right side, with edematous nasopharynx(figure-2), otoscopic examination shows bilateral intact tympanic membrane. On further evaluation routine investigation were found to be normal, FNAC of neck swelling showed differential diagnosis of Kimura's disease, Angiolymphoid, Angioimmunoblastic lymphadenopathy, Hodgkin's disease and suggested biopsy for confirmation. Biopsy of neck node was done under local anesthesia and histopathology confirmed Kimura's disease (figure 5-6). Patient was treated conservatively with iv steroids, iv antibiotics, & oral antihistamines and was discharged with oral steroid & antihistamine. Patient's condition improved with absent neck swelling and no history of recurrence of neck swelling on follow up till date .

III. Discussion:

Kimura's disease is a chronic angiolymphoid proliferative disorder. This disease has male predominance and occurs mostly during the second and third decades of life, endemically in Asia, especially in Japan and China(2–4). As a clinical manifestation, asymptomatic unilateral soft-tissue swellings, such as salivary glands and local lymph nodes, are most common. Other less common involved areas include the axillary, inguinal, and popliteal regions(1). On the other hand, auricular involvement has been reported as a rare event. Kase et al(2) reported only one case involving mainly the auricle among 130 patients with this disease. Bilateral auricular involvement is thought to be extremely rare, and no such case has been reported in the English-language scientific literature. The histopathology findings of this disease consist of hyperplasia of germinal centers and infiltrations of polykaryocytes, eosinophils, blood vessels, and fibrotic changes. These findings of Kimura's disease may mimic angiolymphoid hyperplasia with eosinophilia, but these diseases are thought to be different entities (5). The cause of Kimura's disease remains unknown but is thought to be related to allergic reaction, because the patients often have eosinophilia and high serum immunoglobulin E. Complications such as atopic dermatitis, allergic rhinitis, asthma, and urticaria occur among patients with Kimura's disease, but we cannot find any report of the incidence of the complications. Moreover, the

histopathology findings may support this opinion. In this case, eosinophilia and itchiness were observed. Three therapies are predominantly chosen: corticosteroid therapy, radiation therapy, and surgical resection. Recurrence is common, however, especially when only resection is used, and there has been no definitive treatment to cure this disease(2). We think the initial treatment should be conservative, such as corticosteroid therapy. If no effect is achieved, surgical or radiation therapy can then be considered. The recurrence rate is lower when two or more therapies.

There are many reports concerning CT and MR imaging findings of Kimura's disease. On CT scans, irregularly shaped subcutaneous mass or masses and swelling of the salivary glands are typical findings(6). On contrast-enhanced CT scans, enlarged lymph nodes are enhanced homogeneously, although the salivary glands are enhanced heterogeneously(7, 8). The masses are iso-or hypo intense on T1-weighted MR images and are hyper intense on T2-weighted MR images, compared with the parotid gland(2, 8). Nonetheless, there is a case report showing low signal intensity compared with normal parotid gland on T2-weighted images because of the fibrotic change(4). Thus, the MR findings of Kimura's disease are variable and are thought to depend on vascularity and fibrosis. The findings of CT and MR imaging in our case were compatible with Kimura's disease, except for bilateral involvement. Many diseases involve bilateral auricles or surrounding tissue near the parotid glands. Differential diagnoses include parotid tumors, malignant lymphomas, neurofibromatosis, malignant tumors originated from cutaneous or subcutaneous tissues, metastatic tumors, and inflammatory diseases. Simultaneous Occurrence of tumors in both parotid glands has been reported. These include Warthin's tumor, acinic cell carcinoma, and oncocytoma (9– 11). These parotid tumors are usually encapsulated and are almost limited to the parotid gland, although the lymph nodes of the periparotid region can be involved. Lymphoma or metastatic lymphadenopathies may involve the neck bilaterally (12, 13). Long clinical courses, however, as in Kimura's disease, are unusual. Among inflammatory lesions, tuberculosis and acquired immunodeficiency syndrome should be ruled out. On imaging studies, tuberculosis may show bilateral cervical lymphadenopathies, which tend to have a ring-like contrast enhancement. Bilateral parotid enlargement occurred in patients who had tested HIV-positive and were known to have bilateral parotid cystic lesions(14). Nevertheless, we cannot find any report of Kimura's disease accompanying a ring-like contrast enhancement or cystic components. Although there are many diseases that are associated with bilateral cervical nodular swellings, Kimura's disease should be included in the differential diagnosis. We think that uncommon presentation of kimura's disease in elderly female makes it difficult to diagnose this disease by clinical presentation however careful clinical history, clinical examination gives clue regarding the disease, but the disease is always confirmed by histopathological examination & needs adequate medical treatment & long term follow up to prevent the recurrence of the kimura's disease.

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Figure 1- neck swelling level III LN involvement



Figure 2: nasal endoscopy (edematous & congested nasopharynx)



Figure 3 direct laryngoscopy (edematous epiglottis with edematous bilateral aryepiglottic fold)



Figure 4: direct laryngoscope

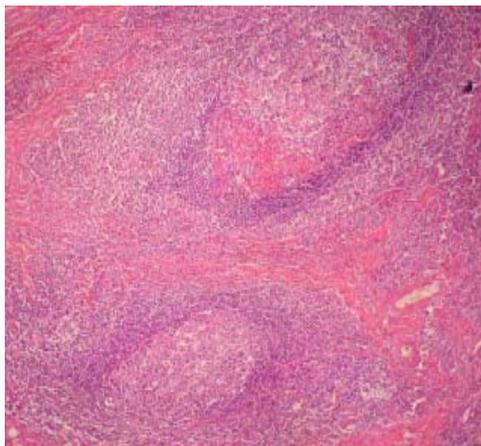


Figure 5: HPE (10X)

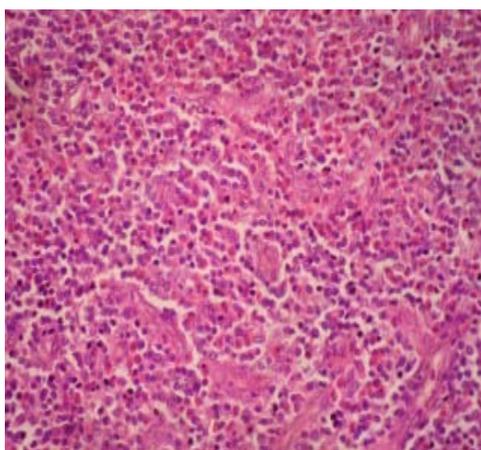


Figure 6: HPE (100X)